Complete Midline Cleft of Lower Lip, Mandible, Tongue, Floor of Mouth with Neck Contracture: A Case Report and Review of Literature

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Abstract

Midline cleft of the lower lip and mandible is an extremely rare condition. Since 1819, when the first case was reported by Couronne, fewer than 80 cases have been described in the world literature so far. The cleft has also been described as facial cleft no. 30 by Paul Tessier. The condition varies in severity from a mild variety in which there is a submucous cleft and notching in the lower lip to a severe variety, involving the tongue, floor of the mouth, mandible, absent hyoid, atrophic neck muscles, and sternum. In this case report, a female child having complete midline cleft of the lower lip and mandible, with bifid tongue stuck to the floor of the mouth, absent hyoid bone and flexion contracture band extending from the confluence of the tip of the tongue, floor of the mouth, cleft mandible to the manubrium sterni is described, with special emphasis on surgical planning and management.

Keywords

- midline mandible cleft
- Tessier no. 30 facial cleft
- surgical management
- bifid tongue stuck with floor of mouth
- flexion contracture band of the neck

Facial clefts involving the upper lip and maxilla are commonly encountered in clinical practice. However, midline clefts of the lower lip and mandible are extremely rare. They are considered to be the result of failure of mesodermal penetration and merging of paired mandibular processes.[1–7]

The median cleft of the lower jaw was first described by Couronne in 1819.[8] Since then, only 80 cases have been reported so far. In 1976, Paul Tessier introduced the number system for craniofacial clefts wherein the mandible midline cleft was notified as no. 30. They are considered as caudal extension of no. 14 cranial and no.0 facial cleft. No. 30 cleft is characterized by mandibular cleft, intermandibular dysplasia, and midline branchiogenic syndrome.[6,9–11]

Case Report

The father of the child (D. N.) contacted the author on the Internet and provided details of the baby with clinical photographs. The child was born through cesarean section after full term, was crying excessively, and unable to take feeds properly (►**Fig. 1**).

The upper lip and nose were normal. The lower lip was split in the midline and the tongue had a deep furrow in midline (►**Figs. 2** and **3**). The tip of the tongue was stuck to the cleft margins of the lower lip and the mandibular cleft ends, which were mobile under the mucosa and floor of the mouth. There was significant ankyloglossia; however, muscle movements were seen under the mucosa. There was excessive drooling of saliva and excoriation of skin over the neck (►**Fig. 4**). There was a flexion contracture band extending from the midline cleft up to the manubrium sterni (►**Fig. 5**). X-ray showed the midline mandibular cleft and hypoplastic hyoid bone (►**Figs. 6** and **7**).

Complete workup was done to rule out any associated anomalies. Initially, the child was on nasogastric feeds and managed by pediatrician. Subsequently after a month, oral feeding was established by spoon. The child gradually picked up weight and was taken up for surgery around 11 months of age.

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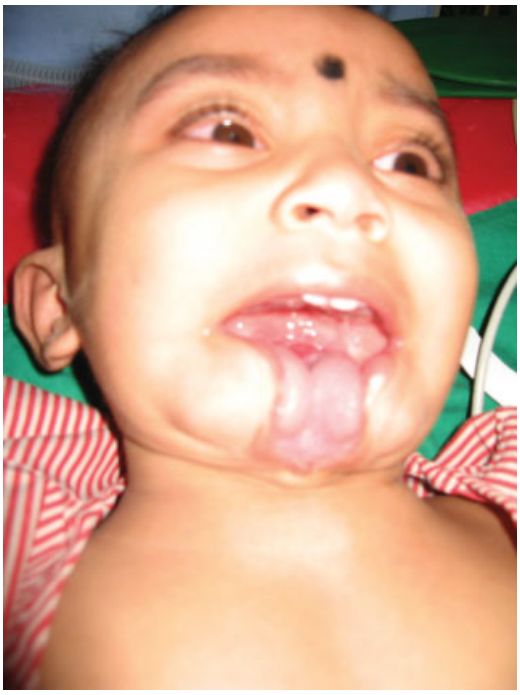

Fig. 1 Preop.

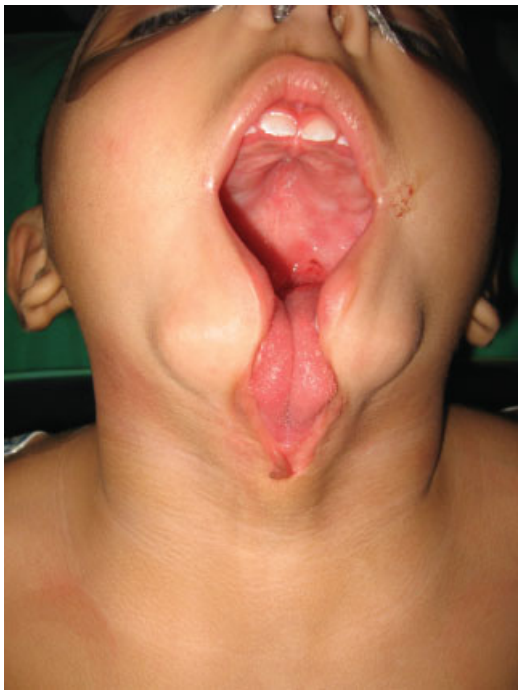

Fig. 3 Bifid chin, tongue stuck to floor of mouth.

Anesthesia Considerations

Considering the general condition of the child, it was decided to do an elective tracheostomy preceding the surgery. Under general anesthesia, an elective preliminary tracheostomy was done using no. 3 Portex tracheostomy tube (Smiths Medical ASD, Inc., Keene, NH), which was subsequently removed on 11th postoperative day.

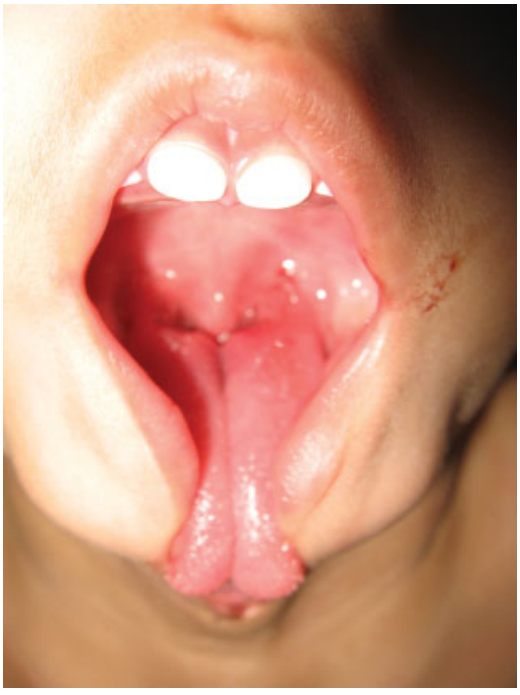

Fig. 2 Child with anomaly.

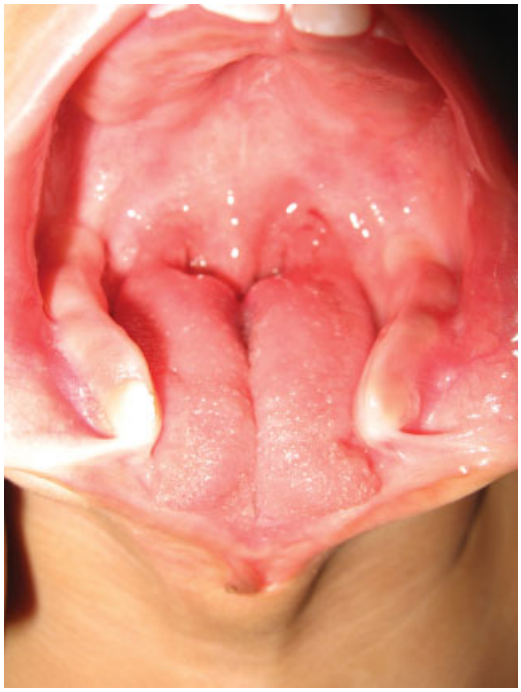

Fig. 4 Mandible cleft with bifid tongue and flexion contracture of neck.

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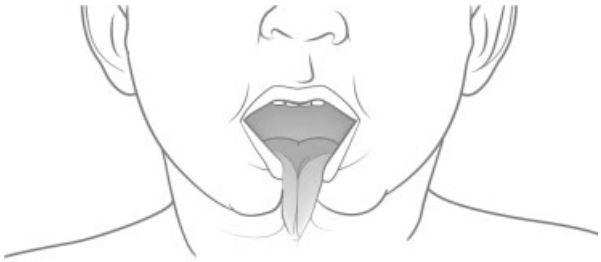

Fig. 5 Illustration showing the extent of anatomical birth defect—midline mandibular cleft.

Surgical Procedure

With elective tracheostomy in position, the first step was to release the tongue from the floor of the mouth (►**Figs. 8** and **9**). The tip of the tongue was stuck to the sternum as can be seen (►**Fig. 8**). The released tongue was sutured in a multiple Z-plasty closure.

The paravestibular mucoperiosteal flaps were raised next, carefully exposing the adjacent margins of the mandibular cleft. Keeping the teeth root buds in mind, a 26 G stainless steel wire was passed through the drill holes and anchored by twisting. The mucoperiosteal flaps were then approximated to ensure a watertight closure from the newly created floor of the mouth (►**Figs. 10–12**).

The flexion contracture band in the neck extending from the tip of the tongue to manubrium sterni was released by zigzag skin approach, achieving near total extension of the neck.

Postoperative period was uneventful, with good tissue healing. The child is being followed up periodically (►**Figs. 13** and **14**). The dentition appeared normally (►**Fig. 15**). The speech is normal (►**Fig. 16**). Another corrective surgery was done around the age of 3 years, for scar revision of the neck and lower lip. Also, the neck contracture was released again (►**Figs. 17** and **18**). Presently, the child is 5 years old and attending regular school (►**Figs. 19** and **20**).

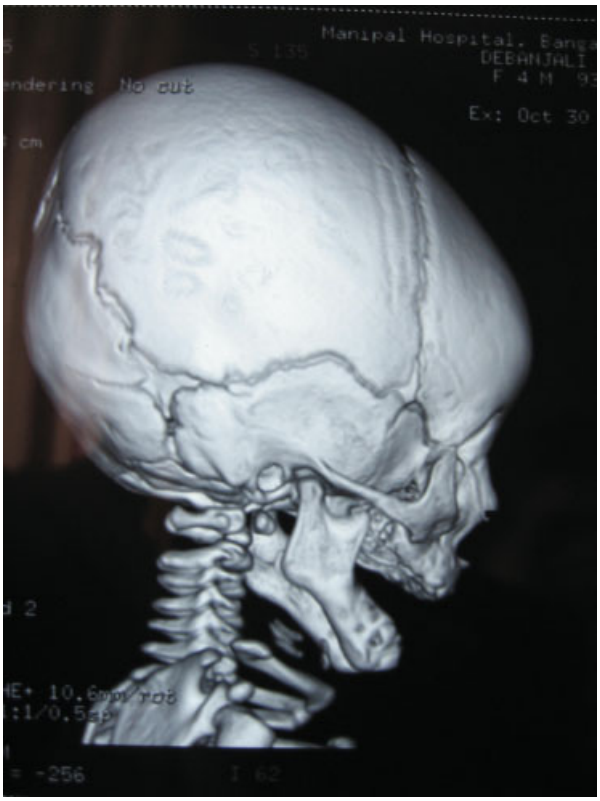

Fig. 7 CT scan showing hypoplastic hyoid bone.

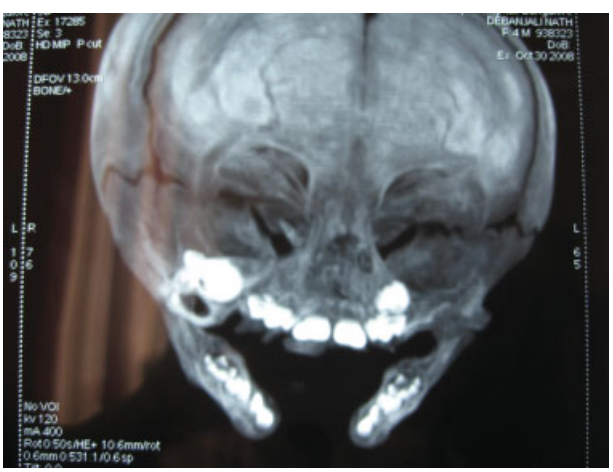

Fig. 6 CT scan showing mandibular cleft.

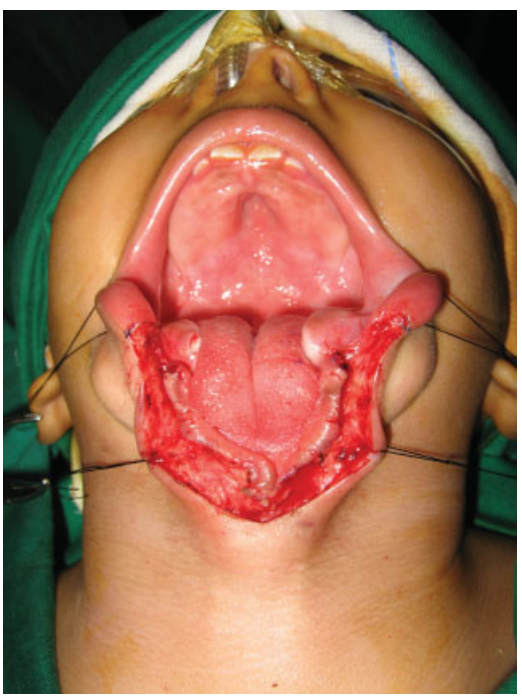

Fig. 8 Release of tongue from floor of mouth.

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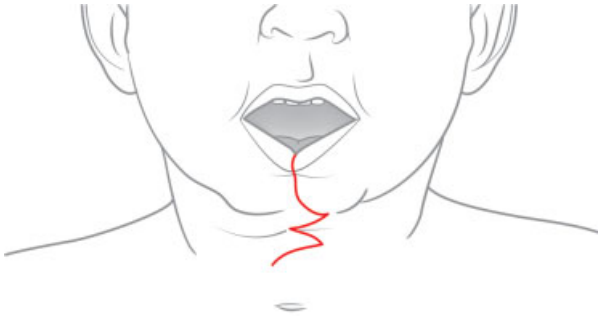

Fig. 9 Planned surgical reconstruction.

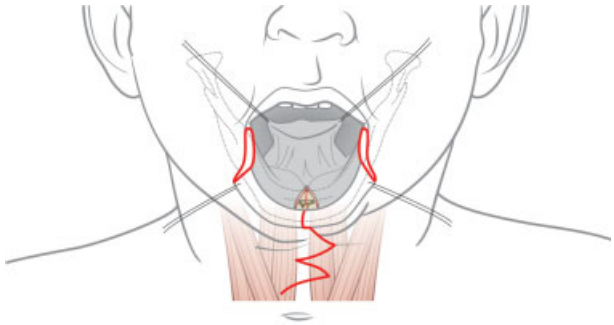

Fig. 11 Details of surgical reconstruction.

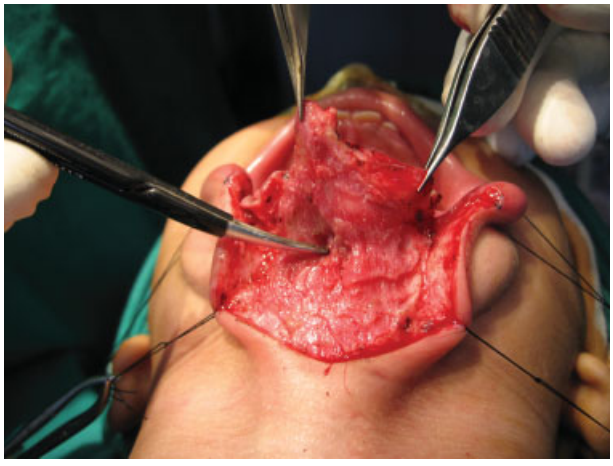

Fig. 10 Total release of base of tongue from floor of mouth.

Discussion

Orofacial developmental abnormalities that involve the upper lip and face are the most common variety. Midline cleft of the mandible (Tessier no. 30) is one of the rarest and almost two centuries after the first case notification, and fewer than 75 to 80 cases have been reported world wide, making it mandatory and worthwhile bringing to light, whenever it occurs.[5,8,10,12,13]

Most authors consider the condition occurs due to a failure of fusion of first branchial arch or improper mesodermal penetration in the midline. This also explains the absence of hyoid, strap muscles, and manubrium sterni in severe cases.[5,6,9,11,13–15]

Hence, the clinical presentation may vary from mild to severe.[6,12] There can be just a notching at the vermillion border.[16] In some cases, the lower lip and chin may be bifid without bony cleft. The anterior aspect of the tongue may be split and attached to mandibular cleft margins with fibrous band. Armstrong reported bifid uvula and ankyloglossia.[10]

The strap muscles of the neck are often atrophic and replaced by dense scar tissue. Monroe reported flexion

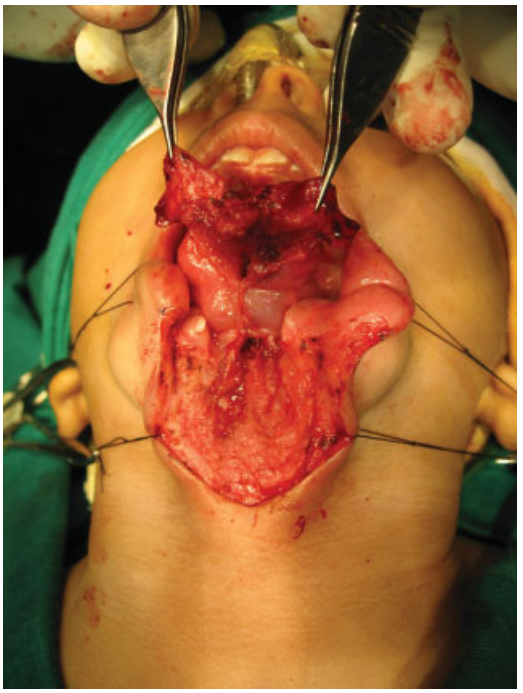

Fig. 12 Reconstruction of tongue and floor of mouth.

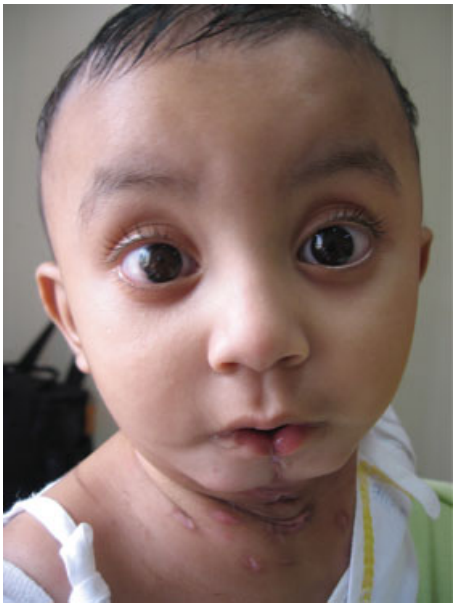

Fig. 13 Early post-op.

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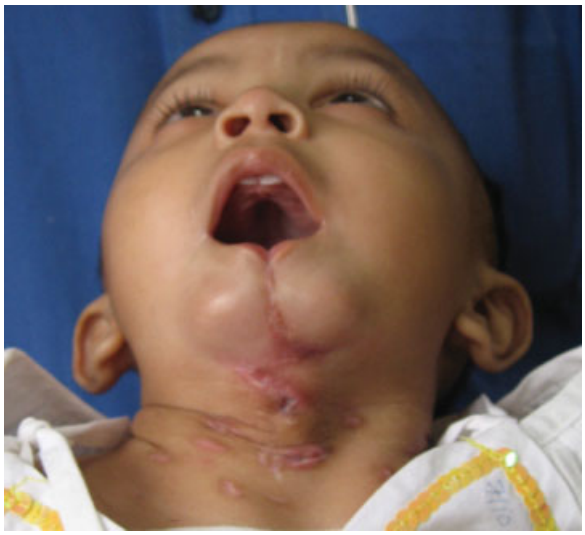

Fig. 14 Early post-op showing good neck.

contracture of the neck.[5] These fibrous bands act as severe flexion contracture bands causing inability to extend the neck, making the child adopt an awkward upward glance, as was evident in the patient.

Other associated facial anomalies such as widened interclavicular space; bifid or absent manubrium; presternal skin tags; cleft lip and palate; mucous pits of lower lip; hemifacial microsomia; and angular or midline dermoid cyst of nose, eye, and ear can exist. Various hand and foot anomalies and ventricular septal defect have also been reported.[5,6,12,13,16–19]

Rana et al have noted ectopic salivary gland on the dorsum of tongue.[20] Pujar and others have reported foregut duplication cyst in the floor of the mouth along with midline mandibular cleft.[21] Surendran and Varghese have published a case of midline mandibular cleft associated with midline dermoid in the neck.[22] Lack of differentiation during late embryonic period may lead to these features.[15] Subramani et al have noted a familial pattern in which mother had cleft tongue, ankyloglossia, linear band in the midline of palate, and daughter was born with cleft tongue, soft palate, ankyloglossia, midline notching of upper lip, and facial ectodermal dysplasia.[23]

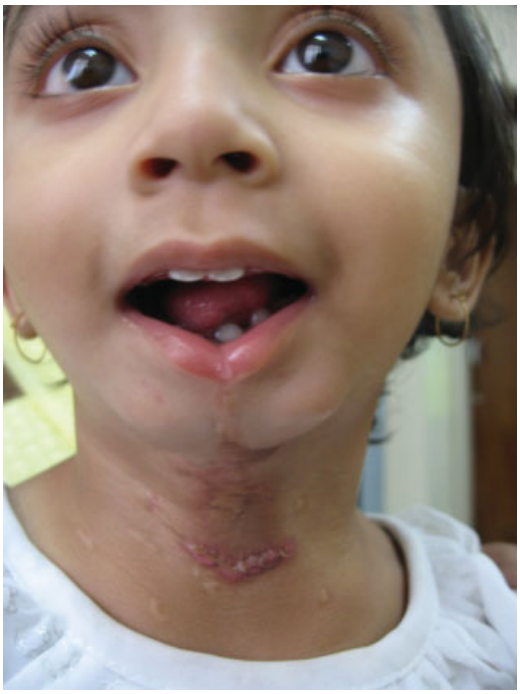

Fig. 16 Child showing good mouth opening and tongue mobility with good speech.

This patient represented the severe form of midline mandibular cleft (Tessier no. 30) having bifid tongue with total ankyloglossia, absent floor of mouth, hypoplastic hyoid, hypoplastic strap muscles of the neck, cleft of lower lip, and an extreme flexion contracture of the neck with the midline fibrous band running from tip of the tongue to manubrium, throwing up many challenges in planning and execution of surgical treatment.

Staged reconstruction, as advocated by Armstrong and Waterhouse, is the most commonly followed surgical protocol in the world.[10] Reconstruction of mandible is done with rib, iliac bone graft, along with titanium or bioresorbable mini-

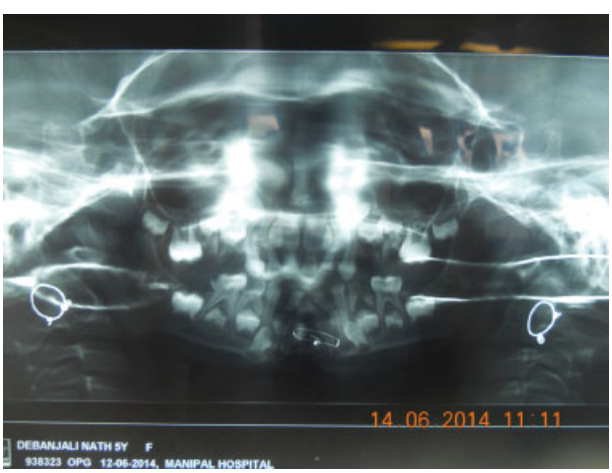

Fig. 15 Fixation of mandibular cleft margins by interosseous wiring.

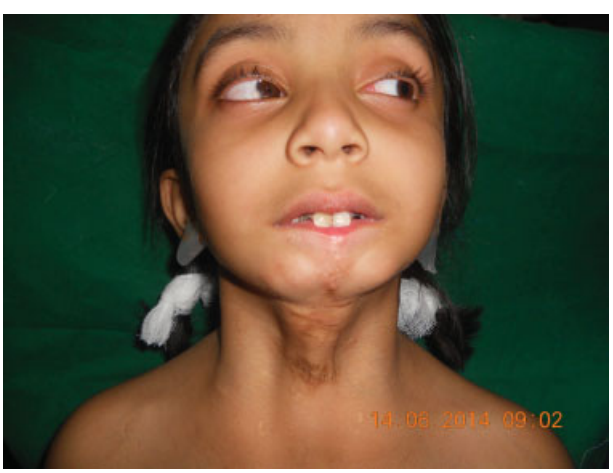

Fig. 17 Follow-up at 3 years.

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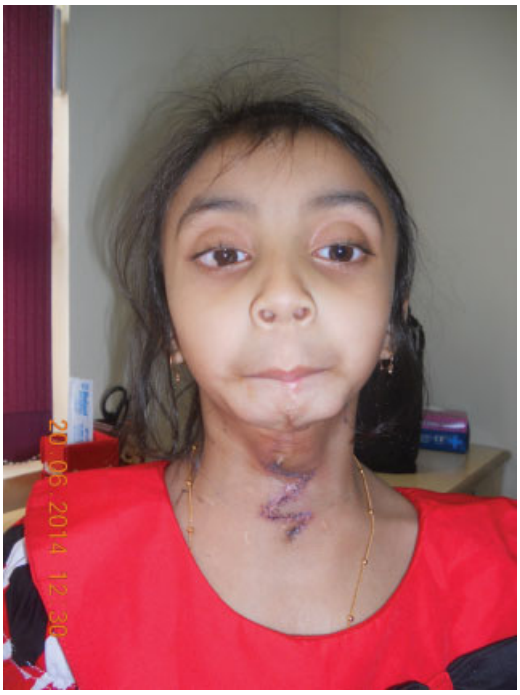

Fig. 18 Serial Z-plasty release of neck flexion contracture.

Fig. 20 Full functional recovery at 5 years follow-up.

plates and mini-screws.[24] Seyhan and Kýlýnr freshened the edges of the mobile mandible segments and fixed them with stainless steel wires in a 10-month-old baby with good result.[18] We used the same procedure in this patient.

Armstrong and Waterhouse also suggested that bony reconstruction should be done preferably after eruption of secondary dentition to lessen the damage to growing tooth buds.[10] In general, if there is no gap between the cleft segments, reconstruction can be deferred till 10 years. If the cleft mandible segments are freely mobile, as in this case, early reconstruction becomes mandatory.[24–27] We decided to stabilize the mobile segments with interosseous stainless steel wiring after careful freshening of the cleft margins, bearing in mind a definitive mandibular reconstruction at a later date.[10,28,29] The latest follow-up of the child, who is now 5 years old, shows a good extension of the neck, stable mandible with good occlusion, and good speech (►**Figs. 19** to **21**).

The child underwent additional surgeries in the form of corrective surgery for the notching of lower lip from the primary lip repair and excision of the atrophic muscles, scar tissue in the neck by serial Z-plasty to achieve a satisfactory mentosternal angle (►**Figs. 17** and **18**).

In one of the largest retrospective analyses reported by Renato Da Silva Frietas and others, 17 patients ranging in age from 1 month to 30 years underwent various staged reconstructive surgeries. Only four patients in their series were similar to this child in severity of presentation. They performed early surgery in two patients for sleep apnea and feeding, using distraction osteogenesis to achieve better tongue position. They used Z-plasty of mucosa, muscle repair,

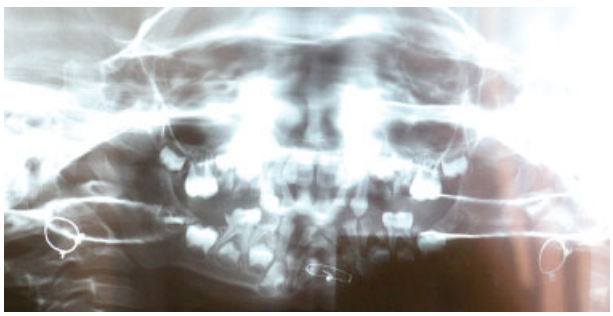

Fig. 19 Appearance of normal dentition and good bony stability at 5 years follow-up.

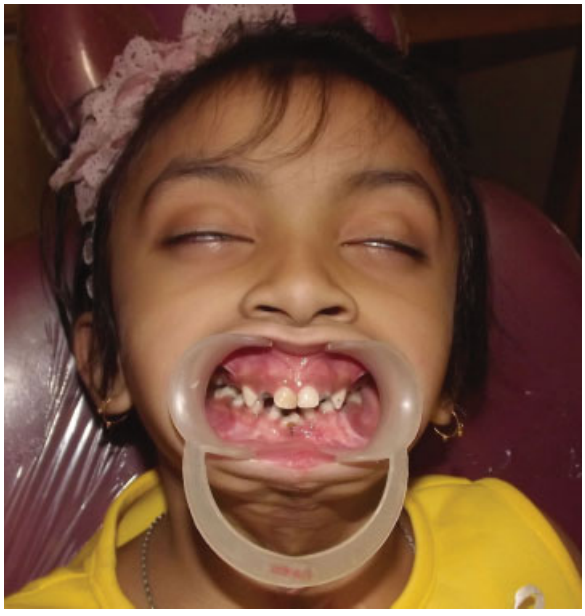

Fig. 21 Dental occlusion at latest follow-up at 5 years.

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and straight line closure for the skin in a few patients. They also recommended early bone grafting to establish the mandibular continuity.[15]

Ishii et al have reported a 17-year follow-up of a patient with midline mandibular cleft of moderately severe grade. The multiple surgeries included primary repair, release of tongue, repair of floor of mouth, and vestibuloplasty at 5 months of age. They performed bone cleft reconstruction in stages—mandibular fixation at 6 years of age followed by definitive bone graft from iliac crest 1 year later. At nine years, all plates were removed. At 11 years, a vestibuloplasty was performed, and at 3 years, augmentation of the mid-mandibular region by onlay iliac bone graft was done. At 16 years, the contracture in submental region was excised using a full-thickness skin graft.[29] We have planned a similar follow-up and surgical protocol for this patient.

Oostrom and others stated that careful osteosynthesis at the inferior border of the mandible will not harm tooth buds. Osteosynthesis may be necessary for the proper development of the teeth. Also, condylar cartilaginous growth is usually dependent on functional stimulation of masticatory activity. Therefore, it appears logical to normalize mandibular function in an early stage of life.[6]

In this patient, the lower border of the cleft mandible segments was stabilized by placement of an interosseous stainless steel wire in the first corrective surgery at the age of 11 months. The procedure is similar to the one described by Seyhan and Kýlýnr.[18] It has held the mandibular cleft segments in good position, dental occlusion, and allowed the baby to have a normal mastication and speech. The latest follow-up of the child, who is now 5 years old, shows a good extension of the neck, stable mandible with good occlusion, and good speech (►**Figs. 19** to **21**).

Conclusion

Midline mandibular cleft and associated clinical anomalies are extremely rare congenital facial deformities. This patient had severe flexion contracture of the neck, total ankyloglossia with absent floor of the mouth, and hypoplastic neck muscles. A clear plan of management, successful surgical correction, and long-term follow-up of the patient, along with review of literature, has been documented in this study.

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